INDIANA
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NEWSLETTER

Indiana State Department of Health

Epidemiology Resource Center
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Transmissible Spongiform Encephalopathy (TSE): History and New Concerns

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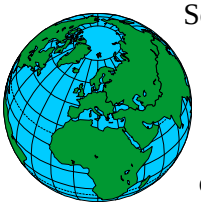
The transmissible spongiform encephalopathies are a group of diseases that cause neurological dysfunction in animals and people. Clinical characteristics of these diseases are loss of motor control, dementia, wasting, paralysis, and eventually death. They have generated a great deal of interest and concern in the last 10-15 years because of the recent identification of a TSE in British cattle (bovine spongiform encephalopathy, or BSE) and its linking in 1996 with new variant Creutzfeldt-Jakob Disease (nvCJD) in several young adults in the United Kingdom. Diagnosis is made at autopsy by the histological changes in the brain that makes the brain tissue appear sponge like.

Animal species in which a TSE has been identified include sheep and goats (scrapie), cattle (bovine spongiform encephalopathy), mink (transmissible mink encephalopathy), mule deer, white-tailed deer and elk (chronic wasting disease), and humans (Creutzfeldt-Jakob Disease, Gerstmann-Straussler-Scheinker syndrome, Fatal Familial Insomnia, Kuru, and Alpers syndrome).

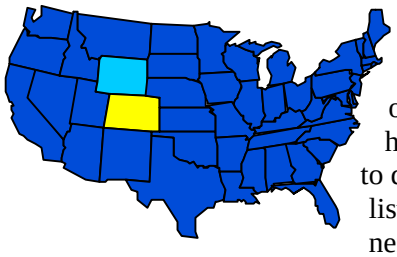
These diseases are thought to be caused by a controversial agent that does not contain any genetic material (DNA or RNA), but consists only of protein. The agent has both an infectious and a hereditary component. There is some evidence that the disease agent is a modified form of a normal cellular protein involved in synaptic function. The normal protein is protease sensitive. The modified form is relatively resistance to protease and accumulates in the cytoplasmic vesicles of affected individuals. The abnormal protein may also occur as the result of a gene mutation that causes the production of the modified form and is passed to the next generation as an autosomal dominant trait. Experimentally prions from infected animals have been injected to mice and subhuman primates, which in turn developed a TSE. All TSE agents are resistant to heat and normal sterilization practices.



Scrapie is the oldest recognized TSE, having been recognized in Great Britain and other Western European countries for over 250 years. Scrapie was first recognized in the United States in 1947, in a Michigan sheep flock. Scrapie is primarily reported in the Suffolk breed but also reported in other breeds. The name is derived from the behavior of infected sheep to scrape their wool off against fixed objects apparently to relieve itching. Other predominant features of the disease are related to motor control with loss of coordination and gait abnormalities. Transmission is thought to be from ewe to lamb and to other sheep through contact with the placenta

and placental fluids. In laboratory settings, the agent has been transmitted to laboratory animals including monkeys. There is no evidence that scrapie has been transmitted to humans. The USDA has launched an effort to identify infected flocks and eradicate the disease. Indiana has three infected Suffolk flocks. Recently an experimental test has been developed to test asymptomatic animals for presence of the agent.

Transmissible mink encephalopathy was first observed in the United States in 1947 is a rare disease of farm raised mink. Since then, it has been reported in the Canada, Finland, Germany, and the former Soviet Union. Clinical signs of the disease are changes in behavior, incoordination, jerking of hind limbs, circling, compulsive chewing, and clenching of the teeth. Cases were seen at several mink ranches in Wisconsin, Minnesota, and Idaho. Epidemiological studies suggest that contaminated feed is a likely source. The diet at one ranch consisted of "downer cattle"(nonambulatory cattle due to a metabolic disease, broken limbs or a neurological disease), fish, poultry, and cereal. Since 1993 the USDA Animal Plant Health Inspection Service has examined the brains of over 8,400 downer cattle for BSE or other TSE and has not found a single case.



Chronic wasting disease (CWD) was first recognized as a clinical syndrome in 1967 and has continued to occur infrequently in limited areas of Colorado and Wyoming. Since 1981, less than 200 cases have been documented. Cases have occurred in both free ranging animals as well as in farmed elk and other confined herds of deer or elk. The disease is characterized by chronic weight loss leading to death. Behavioral changes such as decreased interactions with other animals, listlessness, repetitive walking in set patterns, and in elk, hyperexcitability and nervousness are seen also. In spite of many opportunities for exposure to other ruminants such as cattle, sheep, and goats, there have been no cases of CWD have been reported in these species. The route or transmission is not known, but is thought to be lateral and maternal.

Bovine spongiform encephalopathy (BSE), better known as "mad cow disease" was first recognized as a central nervous system disease of cattle in the United Kingdom in 1986. It has since been seen in 10 other European countries. Approximately 180,000 cases have occurred worldwide with 95% of those in the United Kingdom. The disease is thought to have been introduced into cattle through feed containing meat and bone meal (from dead sheep) that had not been heated sufficiently to destroy the prion. The disease is referred to as "mad cow disease" because of afflicted cow's clinical presentation of behavior changes of nervousness or aggression. Other clinical features are abnormal posture, incoordination and difficulty in rising, decreased milk production, and loss of body condition in spite of continued appetite. The incubation range is six to eight years and following onset of clinical signs, animals die in 2 weeks to 6 months. In 1988, the epidemiology of the outbreak was sufficiently understood that the United Kingdom banned the feeding of ruminant proteins to other ruminants and instituted other steps to eliminate BSE in cattle. The number of cattle cases in the United Kingdom peaked in 1993 and continues to decline.

In March 1996, it was announced that 10 cases of new variant Creutzfeldt-Jakob Disease (nvCJD) had been diagnosed in 1994 and 1995. This new CJD differentiated from classical CJD in several ways. Affected individuals were younger (average age 28 versus 60+); the course of the disease was longer (13 months versus 6 months), the electroencephalograph activity was different, and there was a different brain histopathology, yet recognizable as CJD. Recently reported research in mice supports the theory that BSE and nvCJD are caused by the same agent.

The U.S. Department of Agriculture (USDA) has taken several steps to ensure that BSE does not occur in the United States. The use of ruminants in meat and bone meal was stopped. Importation of cattle from the United Kingdom was stopped and those animals that had been imported were traced and identified to ensure they did not go into the food chain. Most recently, the USDA has been attempting to destroy a herd of sheep imported from Belgium that may be infected with BSE. The farm and its animals have been under quarantine to ensure than none of the animals enters the food chain.

Creutzfeldt-Jakob Disease (CJD) was first described in 1920 by a German neurologist named Creutzfeldt followed by the description of four more cases by A. Jakob the following year. Clinically CJD is a profound dementia with ataxia and diffuse myoclonic jerking. This disease is uncommon, generally occurring in late middle age at a rate of one case per million population worldwide. The disease occurs most frequently as a sporadic event, but has been transmitted iatrogenically by the use of dura mater grafts, corneal transplants, human growth hormone, and brain depth electrodes. A familial occurrence has been recognized but the pathway of transmission is not known.

Since the identification of nvCJD in the United Kingdom, increased attention has been paid to CJD in the United States. The Centers for Disease Control and Prevention (CDC) is studying the 1979-1997 nationwide CJD deaths. During that time period, 4,471 CJD deaths were identified. The average age-adjusted death rate was 0.97 deaths/million persons. Whites accounted for 94.9% of the deaths and the median age was 68 years (64% of the nvCJD deaths in Europe died at less than 30 years of age). Males had a slightly higher death rate than females. Clinical and/or neuropathology records for 77 of 87 patients who died of CJD younger than 55 years of age revealed no evidence of nvCJD. Brain tissue from 269 suspected CJD deaths did identify pathology distinctive nvCJD. The rate of CJD deaths in the United States has remained stable from 1979 to 1997 and no nvCJD deaths have been identified.

Indiana incidence of CJD is very similar to the national statistics with the exception that 60% of the cases were females. Figures 1 and 2 provide a graphic description of the Indiana CJD deaths by year and age. The mean and median age of cases was 68.8 years, and 80% of the cases were 60 years of age or older (more than 50% of the vCJD deaths in the United Kingdom were less than 30 years of age). Since 1990, the number of cases has ranged from 2 to 9, but the mean number has been 5.1 cases/year. Indiana's death rate (0.87cases/million population) is slightly lower than the national average (0.97cases/million population). From all appearances Indiana's CJD deaths are typical of CJD. Health care workers should be aware of the potential for iatrogenic transmission, and other citizens should be aware that CJD is not easily transmitted and is not communicable in ways normally associated with infectious diseases.

Figure 1.

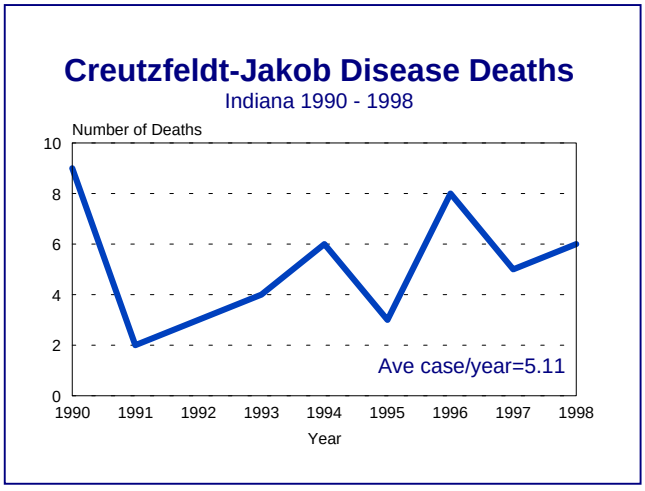
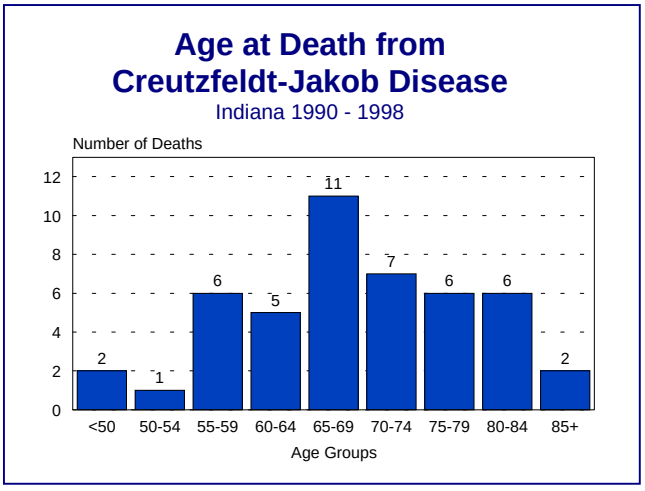


Figure 2.



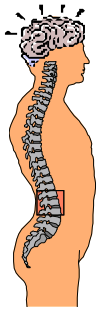
Another human spongiform encephalopathy known as Kuru was identified in the early 1900 in a New Guinea tribe that practiced mortuary ritual acts of cannibalism. With the stopping of the cannibalism practices, Kuru has disappeared.

References:

1. Principles and Practice of Infectious Diseases, Mandell, Douglas, Bennet, 3rd Ed. Churchill and Livingstone, 1990.
2. The Prion Disease, Scientific American, Jan 1995
3. Creutzfeldt-Jacob Disease Surveillance in the United States, Gibbons, Belay, Holman, Shahriare, Schonberger. Abstract, International conference on Emerging Infectious Diseases 2000.
4. http://www.as.ua.edu/ant/bindon/ant570/Papers/McGrath/McGrath.htm, Kuru: The Dynamics of a Prion Disease,
5. http://www.aphis.usda.gov/oa/pubs/fstme.html, Transmissible Mink Encephalopathy
6. http://www.aphis.usda.gov/oa/pubs/fsscapie.htm, Scrapie
7. http://www.aphis.usda.gov/oa/bse, Bovine Spongiform Encephalopathy (BSE)
8. http:/www-micro.msb.le.ac.uk/335/Prions.html, Prion Diseases
9. Creutzfeldt-Jacob disease: Recommendations for infection control, Victoria McGreevy Steelman, R.N., American Journal of Infection Control, Vol. 22. No.5, Oct 94, 312-318.

Meningitis Update

Stephanie Fang, RN, BSN
ISDH Communicable Disease Division



Meningitis is an infection of the fluid of the brain and spinal cord. The usual symptoms of meningitis include fever, headache, and neck stiffness, which may develop in as little as two hours. Confusion, rash, nausea, and vomiting may also be present.

Each year, the reported incidence of meningitis in the United States is 2.2 cases per 100,000, with about one-third of all cases occurring in children under 5 years of age. Meningitis is usually caused by bacterial or viral infections. It is important to determine the causative organism as the public health implications vary significantly.

Bacterial Meningitis

Before the 1990s, *Haemophilus influenzae type B* (Hib) was the leading cause of bacterial meningitis. Initiation of Hib vaccine into the routine childhood vaccination schedule has greatly reduced the incidence of disease due to *H. influenzae*. Currently, the two leading causes of bacterial meningitis are *Streptococcus pneumoniae* and *Neisseria meningitidis.*

Bacterial meningitis is usually more severe than viral and may progress to seizures, shock, coma, and death. Prompt medical evaluation and treatment is critical. Cultures of blood, cerebrospinal fluid, synovial fluid, pleural fluid, pericardial fluid, petechial, or purpuric lesions will confirm diagnosis.

Bacterial meningitis can be contagious. Infection is spread through direct contact with nasopharyngeal secretions. Disease risk can be measured as follows:

High risk

- household contacts
- child care or nursery school contacts
- direct exposure to nasal/oral secretions through kissing or sharing toothbrushes or eating utensils

Low risk

- casual contact – no direct exposure to nasal/oral secretions
- indirect contact – only contact is with high risk contact

Early diagnosis is essential for antibiotic treatment to be effective. Some agents of bacterial meningitis exhibit antibiotic resistance. When *N. meningitidis* is the cause, chemoprophylaxis is indicated for high-risk contacts.

There are now vaccines for prevention of some types of bacterial meningitis. In addition to the Hib vaccine, pneumococcal and meningococcal vaccines are effective against *S. pneumoniae* and *N. meningitidis* respectively.

On October 20, 1999, the Advisory Committee on Immunization Practices (ACIP) modified its guidelines for the use of the polysaccharide meningococcal vaccine, particularly for college freshmen who live in dormitories, a group found to be at a modestly increased risk of meningococcal disease relative to other persons their age.

The ACIP recommendation states that medical providers who provide care to these students give information about the disease and the benefits of vaccination to the students and their parents. In addition, college freshmen who wish to reduce their risk of disease should be given the vaccination or directed to a site where vaccine is available. The full text version can be found at http://www.cdc.gov/epo/mmwr/preview/mmwrhtml/rr4907a2.htm.

While bacterial meningitis has been removed from the reporting rule, many of the organisms that cause bacterial meningitis are still reportable: *H. influenza*, *Listeria*, *N. meningitidis*, *Group A Streptococcus*, *Group B Streptococcus*, and *S. pneumoniae*.

Viral (Aseptic) Meningitis

Viral meningitis is a somewhat common, and rarely fatal, condition. It usually manifests as a clinical syndrome with symptoms of meningeal involvement and, in most cases, recovery is complete. Viral agents can be identified in about half of the isolates tested. CSF findings may include pleocytosis, increased protein, normal sugar, and absence of bacteria.

Aseptic meningitis is a reportable disease and there are seasonal variations in case incidence. However, actual incidence is unknown due to the difficulty in isolating specific virus types. Enteroviruses (i.e., coxsackievirus and echovirus) are responsible for most cases with known etiology in the United States.

Precautions and preventive measures are dependent on the etiology, which is often not known until after recovery. Therefore, no special precautions are recommended beyond routine sanitary practices. In addition, there is no treatment or vaccine for most causes of viral meningitis.

For more information about bacterial and viral meningitis, visit the following web sites:

Centers for Disease Control and Prevention:	www.cdc.gov
Indiana State Department of Health:	www.state.in.us/isdh/

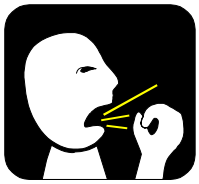
Pertussis Reports Increasing in August

Wayne Staggs, MS
ISDH Epidemiology Resource Center

Pertussis reports in Indiana have increased during August, with 31 cases being reported between August 1-17. All 31 cases were in various stages of investigation at the time this newsletter was published. Eighteen of the cases are residents of Allen County. Although none of the Allen County cases have been cultured confirmed, 13 do meet the clinical case definition for pertussis. Six of the Allen County cases, four of which meet the clinical case definition, attend the same day care center. Other counties reporting pertussis cases since August 1 include DeKalb (6), Vigo (2), Lake (2), Madison (1), Clark (1), and Jackson (1).

In a typical year in Indiana, approximately 50% of pertussis cases that are eventually reported to the MMWR have cough onsets occurring in the three-month period from July - September. **The increased reporting statewide and in Allen County serves as a reminder that all suspected cases should be reported to the local health department.** A follow-up investigation of each reported case is conducted by a state immunization investigator. It is preferable that physicians obtain laboratory specimens for DFA and culture analysis followed by treatment of the patient and all household contacts with appropriate antibiotic prophylaxis. If laboratory specimens cannot be obtained, antibiotic treatment should begin immediately. Persons are considered non-contagious after 5 days of antibiotic treatment, but should complete the entire course of antibiotics.

Pertussis is the most frequently reported vaccine-preventable disease reported among children less than five years of age. As of August 18, 42 cases of pertussis have been reported to the MMWR (does not include cases under investigation), of which 22 (54.7%) were under the age of five. However, due to waning immunity following either natural infection or vaccination, pertussis can affect persons of any age and is increasingly recognized in older children and adults. Of the 42 case reported thus far in 2000, 11 (26.1%) are among persons 20 years and older. **Physicians should include pertussis in the differential diagnosis of cough illness in persons of all ages, regardless of their immunization status.** Until approved booster vaccination for pertussis is available to protect older children and adults, prompt diagnosis and treatment of cases and prophylaxis of contacts are the primary methods of limiting transmission. Further information on pertussis control can be found in the ISDH Recommended Pertussis Control Measures. (To obtain a copy, contact your Immunization Field Representative or call (317) 233-7112.)



Pertussis is an acute respiratory infection caused by the bacteria *Bordetella pertussis.* The illness begins with an onset of mild upper respiratory symptoms, which progresses after about a week to a paroxysmal cough. Coughing may last several weeks, even following appropriate antibiotic therapy. Fever is usually minimal or absent. Transmission is primarily through direct contact with respiratory secretions of an infected person. The incubation period is usually 7-10 days, but can extend to 21 days on rare occasions. The clinical case definition for pertussis is a cough of 14 days or longer and one of the following: paroxysmal cough, whoop, or vomiting after coughing.

Conferences and Seminars

Fall Immunization Award/Educational Conferences Coming Up in October

October 25
Jasper Holiday Inn
Jasper, Indiana

Welcome:
Dr. Thomas Gootee
Health Officer
Dubois County Health Dept.

Keynote Speaker:
Dr. Richard Clover
Prof., Family & Community Med.
University of Louisville, KY

October 26
Valle Vista Conference Center
Greenwood, Indiana

Welcome:
Dr. Richard Feldman
IN State Health Commissioner

Keynote Speaker:
Dr. John Gaebler
Assoc. Prof., Pediatrics
Indiana Univ./Purdue Univ.

October 27
South Bend City Centre Holiday Inn
South Bend, Indiana

Welcome:
TBA

Keynote Speaker:
Phillip Hosbach IV
Aventis Pasteur

For more information, please call Pat Schwer at the Indiana State Department of Health at (317) 233-7704.

Educational Opportunity for Local Health Departments coming up in September:

Satellite Conference on Bioterrorism

Knowledge about the extensive biological weapons programs in other countries and numerous recent Bioterrorism threats have increased the concern regarding the medical management of biological agent casualties on the battlefield overseas and domestically, including Indiana. Therefore, world-renowned experts from the Centers for Disease Control and Prevention (CDC) and the U.S. Army Medical Research Institute for Infectious Diseases (USAMRIID) are co-sponsoring a satellite-based course on Bioterrorism.

During this 3-day, interactive satellite broadcast, participants will learn what the military and civilian medical and public health response should be. Proficiency will be acquired in recognizing that a biological attack has occurred, activating the appropriate agencies and personnel to investigate the event, treating casualties, and preventing the spread of the agent. This program is intended for public health professionals such as local health officers, administrators, environmental health specialists, epidemiologists, and public health nurses involved in disease surveillance and prevention at the local health department level.

Course Registration & Other Information

The satellite broadcast will air:

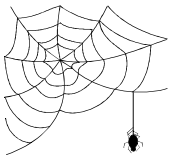
September 26, 27, & 28
12:30-4:30 pm Eastern Daylight Time
OR
11:30-3:30 pm Eastern Standard Time

SEATING IS LIMITED to 15 persons. All sessions will be in the Commissioner's conference room (not the Board room):

Indiana State Department of Health (ISDH)
2 North Meridian Street
Indianapolis, Indiana

Local health department personnel are invited and encouraged to take part in the course. There is no charge for the course.

To register for this satellite broadcast, you MUST register (or have someone with Internet access register you) on the CDC web site at http://www.biomedtraining.org. Continuing education credit is available for a variety of professions. Please read the information on the web site carefully. All registration material, course overview, and other viewing site information is available on the CDC web site. If you have any further questions, please contact Rob Clark of the ISDH Surveillance Investigation Unit, Epidemiology Resource Center, by calling (317) 233-7121 or e-mail rclark@isdh.state.in.us.



Wonderful Wide Web Sites

ISDH Data Reports Available

The ISDH Epidemiology Resource Center has the following data reports and the Indiana Epidemiology Newsletter available on the ISDH Web Page:

http://www.state.in.us/isdh/ (under Data and Statistics)

Indiana Cancer Incidence Report (1990, 95)

Indiana Cancer Mortality Report (1990-1994)

Indiana Health Behavior Risk Factors (1995-96, 97, 98)

Indiana Hospital Consumer Guide (1996)

Indiana Marriage Report (1995, 96, 97)

Indiana Mortality Report (1995, 97)

Indiana Natality Report (1995, 96, 97)

Indiana Natality/Induced Termination of Pregnancy/Marriage Report (1998)

Indiana Report of Diseases of Public Health Interest (1997, 98)

The following site allows access to the web page for any state health department in the United States:

http://www.polsci.wvu.edu/grad/klase/STATEHEALTH/sthlth.html

HIV Disease Summary

Information as of July 31, 2000 (population 5,840,528)

HIV - without AIDS to date:

228	New cases from August 1999 thru July 2000	12-month incidence	3.90 cases/100,000
3,171	Total HIV-positive, without AIDS on July 31, 2000[1]	Point prevalence	54.30 cases/100,000[1]

AIDS cases to date:

326	New AIDS cases from August 1999 thru July 2000	12-month incidence	5.58 cases/100,000
2,566	Total AIDS cases on July 31, 2000[1]	Point prevalence	43.94 cases/100,000[1]
5,923	Total AIDS cases, cumulative (alive and dead)		

[1]Counting only cases alive in July 2000

REPORTED CASES of selected notifiable diseases

Disease	Cases Reported in July		Cumulative Cases Reported through July	
	1999	2000	1999	2000
Campylobacteriosis	73	78	261	260
E. coli O157:H7	12	15	28	40
Giardiasis	62	28	255	243
Hepatitis A	6	4	64	35
Hepatitis B	1	4	27	30
Legionellosis	4	5	23	22
Lyme Disease	2	3	9	10
Meningococcal, invasive	0	6	31	33
Pertussis	15	11	29	38
Rocky Mountain Spotted Fever	6	1	8	1
Salmonellosis	55	41	242	290
Shigellosis	49	152	102	891
Tuberculosis	21	11	81	77
Animal Rabies	0	0	0	0

For information on reporting of communicable diseases in Indiana, call the *ISDH Communicable Disease Division* at (317) 233-7665.

Indiana
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Newsletter

The *Indiana Epidemiology Newsletter* is published by the Indiana State Department of Health to provide epidemiologic information to Indiana health professionals and to the public health community.